Egyptian Journal of Ear, Nose, Throat and Allied Sciences xxx (2016) xxx–xxx

HOSTED BY

ELSEVIER

Contents lists available at ScienceDirect

Egyptian Journal of Ear, Nose, Throat and Allied Sciences

journal homepage: www.ejentas.com

Case report

Juvenile ossifying fibroma of the maxilla

Ben Gamra Ollfa, Romdhane Nadia, Nefzaoui Safa *, Mahjoubi Madiha, Abid Wafa, Hariga Ines, Mbarek Chaouech Chiraz

Habib Thameur Hospital, ENT Department, Tunis, Tunisia

ARTICLE INFO

Article history:
Received 26 June 2016
Accepted 15 September 2016
Available online xxxx

Keywords:
Juvenile ossifying fibroma
Fibro-osseous tumours
Surgery
Recurrence

ABSTRACT

Introduction: Juvenile ossifying fibroma is a rare benign fibro-osseous lesion. It is characterized by the early age of onset, the localization of the tumor, the radiological pattern and a tendency to recurrence. In the light of a clinical case, we review the clinical, histological and radiological features of this tumor.
Case report: We report a case of a nine year old male child who presented with a unilateral swelling of the right middle face with right proptosis. Clinical examination showed a mass filling the right nasal cavity. CT scan and MRI were performed. Biopsy suggests a fibro-osseous lesion. The patient undertook surgery by a degloving approach with complete tumor removal. The histopathological examination revealed the diagnosis of a juvenile ossifying fibroma. No recurrence was noted.
Conclusion: Because of its aggressive and compressive nature, juvenile ossifying fibroma requires an early complete surgical excision. A long-term clinical and radiological surveillance is necessary to prevent recurrence.

1. Introduction

Juvenile Ossifying fibromas (JOF) are benign fibro-osseous tumours characterized by a rapid growth and a high tendency for recurrence.[1] Their diagnosis and management still remains difficult and controversial. Clinical presentation depends on the site of the JOF and may include facial swelling, tooth displacement, nasal obstruction and orbital proptosis.[1]

The recommended treatment is complete surgical excision. A long-term clinical and radiological surveillance is necessary to detect recurrence.[2]

We report a case of juvenile ossifying fibroma involving the ethmoid sinus and the nasal cavity with successful management.

2. Case report

A nine year old male child presented with a unilateral right nasal obstruction with recurrent epistaxis since seven months. He had no family history; however we noted an episod of right acute dacryocystitis treated one year ago.

Clinical examination revealed an unilateral swelling of the right middle face with a marked increase in the intercanthal distance associated with a proptosis (Fig. 1). Intraoral examination was with no abnormalities and the mouth opening was normal. No cervical lymphadenopathy was noted.

On endoscopic nasal examination, a mass lined by normal mucosa filling the right nasal cavity was noted. Ophthalmological examination found no signs of oculomotor paralysis and no loss of visual acuity or visual field abnormalities. Remaining head and neck and systemic examination were without abnormalities.

Computed tomography (CT) of the facial bones demonstrated a well-defined heterogeneous tumor measuring $45 \times 42 \times 32$ mm with diffuse scattered radiopaque foci occupying the ethmoid and the right nasal cavity (Figs. 2–4). This lesion was surrounded by smooth, well-defined cortical bone. It caused displacement of the adjacent inferior and medial orbital plate, the medial wall of the right maxilla and the nasal septum without complete destruction of the bony margins but with a focal osteolysis. On MRI, this lesion has an heterogeneous lowintensity signal on T1-weighted sequences and a lowintensity signal on T2-weighted sequences with intense enhancement in gadolinium injection sequences (Fig. 5 and 6).

A nasal biopsy was performed to confirm the type of the lesion. The results of histopathological examination were consistent with fibro-osseous lesions.

Peer review under responsibility of Egyptian Society of Ear, Nose, Throat and Allied Sciences.
* Corresponding author.
E-mail address: safanefzaoui@gmail.com (N. Safa).

http://dx.doi.org/10.1016/j.ejenta.2016.09.002
2090-0740/

Please cite this article in press as: Ollfa B.G., et al. Juvenile ossifying fibroma of the maxilla. Egypt J Ear Nose Throat Allied Sci (2016), http://dx.doi.org/10.1016/j.ejenta.2016.09.002

Fig. 1. Clinical Frontal view revealing a unilateral swelling of the right middle-face.

Fig. 2. Computed tomography Coronal view bone window setting showing a well defined heterogeneous tumor occupying the maxillary, the ethmoid and the nasal cavity.

Under general anaesthesia, a Degloving approach was made and surgical excision of the tumor was performed. The tumor was completely removed (Fig. 7).

The diagnosis of juvenile trabecular ossifying fibroma was confirmed histopathologically (Fig. 8 and 9).no surgical complications were noted. There were no signs of recurrence on examination and on the follow-up imaging performed one year after the operation (Fig. 10 and 11).

Fig. 3. Computed tomography. Axial view bone window setting showing the tumor with displacement of the nasal septum.

Fig. 4. Computed tomography. Axial view bone window setting showing the displacement of the medial orbital plate.

3. Discussion

Juvenile ossifying fibroma is a benign neoplasm of bone. It is characterized by the early age of onset, the localization of the tumor, the radiological pattern and a tendency to recurrence.[2,3]. Two forms of juvenile ossifying fibromas have now been distinguished: psammomatoid juvenile ossifying fibroma and trabecular juvenile ossifying fibroma.

Trabecular juvenile ossifying fibroma affects children under the age of 15 years in more than 80% of cases.[4] In our case, our patient was a nine year-old child. Both males and females are equally affected.[2,5]

Ossifying fibroma arises in the jaw, facial bones and skull. In contrast, juvenile ossifying fibroma involves most commonly the paranasal sinuses and periorbital bones in 90% of cases.[2–4]

Please cite this article in press as: Ollfa B.G., et al. Juvenile ossifying fibroma of the maxilla. Egypt J Ear Nose Throat Allied Sci (2016), http://dx.doi.org/10.1016/j.ejenta.2016.09.002

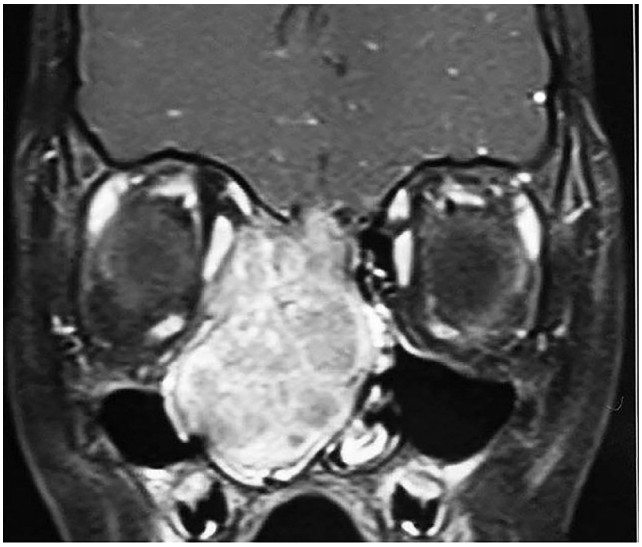

Fig. 5. MRI. Gadolinium-enhanced T1-weighted sequence coronal view: intense enhancement of gadolinium.

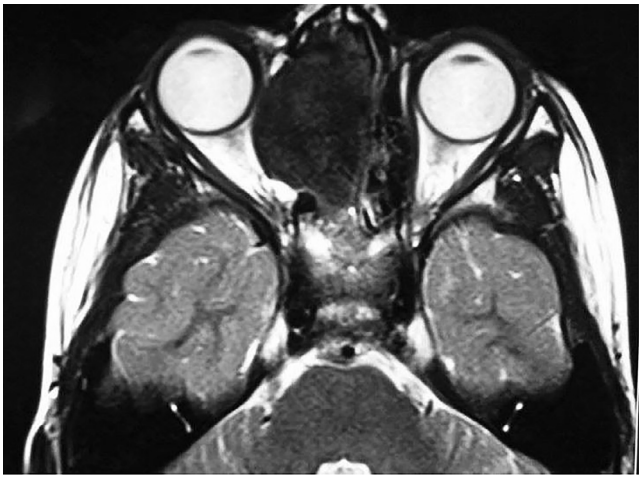

Fig. 6. MRI T2-weighted sequence axial view: lowintensity T2 tumor with extension in the left orbite.

Fig. 7. Operative view of Degloving approach.

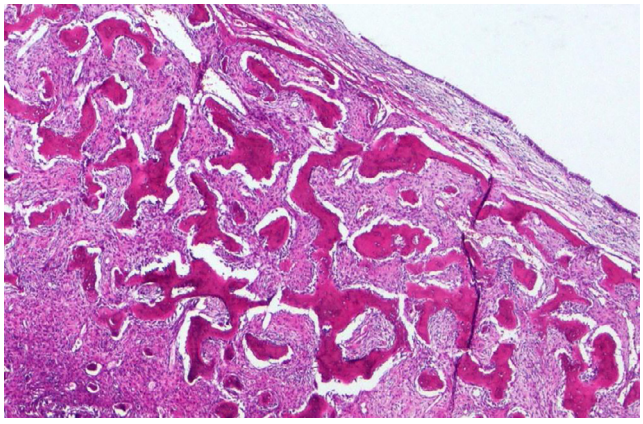

Fig. 8. Microscopic view showing the well circumscribed lesion consisting of a trabecular bone associated with a densely cellular connective tissue.

This rare tumor has a more aggressive behavior than ossifying fibroma.[6] In fact, it tends to erode bones and to extend to adjacent orbital, nasal, and cranial compartments, distorting the face, displacing orbital contents and adjacent structures.

Clinically, it presents as an asymptomatic tumor of aggressive appearance. The symptoms are variable and consist on facial swelling, sinusitis, nasal obstruction, teeth displacement, eye proptosis, visual disturbances, progressive blindness, airway obstruction, and progressive craniofacial deformities.[3,5,7–10]

The local invasion may include pushing and displacement of adjacent bony limits or invasion through osseous delimiting walls with extension into adjacent anatomic compartments.[11,8]

In our case, this tumor was responsible for a displacement of the nasal septum, the inferior and the medial orbital plate and the middle wall of the maxilla.

Histologically, these lesions are always benign, composed of highly vascular and fibroblast-rich connective tissue with calcified substance shaped on bony trabeculae distributed throughout the fibrous stroma. The highly cellular nature of the fibrous matrix reflects the more aggressive behavior of the tumor.[2] Differential diagnoses are mainly other fibro-osseous lesion such as fibrous dysplasia, focal cement-osseous dysplasia.[12,13]

Radiologically, JOF appears as a well circumscribed solitary lesion concentrically expanding, with bone density.[4,9,10,12] The radiolucency of the lesion varies, depending on the maturation stage and amount of calcification[3,14]. The demarcation of the tumor from the surrounding bone is well-defined by a radio-opaque border.[3,12] This aggressive lesion can cause expansion, cortical thinning as well as perforation of surrounding bone.

On MRI, this lesion has a heterogeneous high-intensity signal on T1-weighted sequences and a lowintensity signal on T2-weighted sequences.[3,4] MRI allows defining the extent of the lesion but not clearly its bony component. However, the various histological subtypes of fibro-osseous tumours cannot be distinguished radiologically.

Please cite this article in press as: Ollfa B.G., et al. Juvenile ossifying fibroma of the maxilla. Egypt J Ear Nose Throat Allied Sci (2016), http://dx.doi.org/10.1016/j.ejenta.2016.09.002

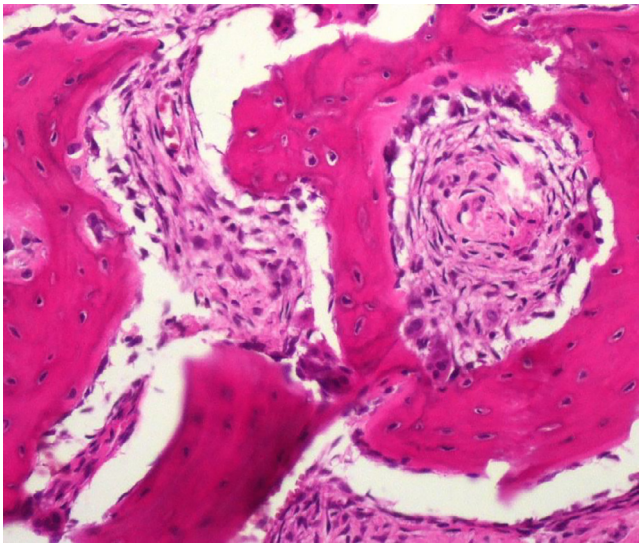

Fig. 9. Microscopic view with high magnification showing a ribbon like osteoid trabeculae lined by osteoblasts and multinucleated giant cells.

Fig. 10. Computed tomography postoperatively axial view: no recurrence.

Surgery is the mainstay of treatment. The reference treatment consists of complete surgical resection.[8]

Some small sinonasal cases can be successfully managed endoscopically.[14] However, an open surgical approach or combined approach (endoscopic and external) is recommended in large tumours that infiltrate sinuses and fronto-nasal bones because it allows complete removal,[2,8] Various external approaches have been used according to the location of the lesion.[7] The tumor mass must be removed down to the level of normal bone with preservation of adjacent vital structures as much as possible.[8]

Immediate reconstruction is not advised because of high recurrence rate. Secondary reconstruction if needed may be undertaken sooner for slow-growing lesions (<1 year), and be delayed for fast-growing lesions (>1 year).[8] We have opted for a degloving approach for our patient to ensure the complete removal of the tumor. We haven't needed any reconstruction.

Fig. 11. Computed tomography postoperatively coronal view: no recurrence.

Radiotherapy is contraindicated because it can cause malignant change.[3] Malignant transformation and metastasis has not been reported but recurrence is common.[3,14]

Local recurrence is usual if the tumor is not completely removed, however some authors evoke the role of dysplastic processes in the bone metabolism.[3] Recurrence rate ranges from 30% to 58% and occurs after period ranging from 6 months to 19 years.[2,3,8,14–16] Evolutive lesions may lead to significant cortical destruction and periosteal elevation with increasing risk of recurrence. The prognosis is considered to be good despite the local invasion.[7,8] For our patient, no recurrence was observed after one year of follow-up.

4. Conclusion

Juvenile ossifying fibroma is described by its aggressive local behavior and high recurrence rate. For these reasons, establish early diagnosis is important and leads to apply the appropriate treatment. Complete surgical resection is the treatment of choice. A long term follow up is recommended to detect recurrences.

Conflict of interest

None.

References

1. Oukabli M, Akhaddar A, Qamouss O. Nasoethmoidal psammomatoid cemento-ossifiying fibroma with intraorbitalextension. *Rev Stomatol Chir Maxillofac*. 2010;111:43–45.
2. Kamal MS, Bahar MAA, Latif MA, Khan MMIU. Juvenile ossifying fibroma of the maxilla: a case report. *AKMMC J*. 2013;4(1):42–44.
3. Chrcanovic BR, Belini FM. An expanded juvenile ossifying fibroma in maxillary sinus: a case report. *J Korean Assoc Oral Maxillofac Surg*. 2011;37:127–132.
4. Breheret R, Jeufroy C, Cassagnau E, Malard O. Juvenile ossifying fibroma of the maxilla. *Eur Ann Otorhinolaryngol Head Neck Dis*. 2011;128:317–320.
5. Pirana S, Zerati F, Voegels R, Maia R. Psammomatoid ossifying fibroma. *Rhinology*. 2003;41:250–252.
6. Hachach-Haram N, Benyon S, Maling S, Joshi N, Grant W, Kirkpatrick WNA. Surgical management of two complex cases of large juvenile orbital ossifying fibroma. *J Plast Reconstr Aesth Surgy*. 2011;64:1661–1664.
7. Choi YC, Jeon EJ, Park YS. Ossifying fibroma arising in the right ethmoid sinus and nasal cavity. *Int J Pediatr Otorhinolaryngol*. 2000;54:159–162.
8. Sarode SC, Sarode GS, Waknis P, Patil A, Jashika M. Juvenile psammomatoid ossifying fibroma: a review. *Oral Oncol*. 2011;47:1110–1116.

Please cite this article in press as: Ollfa B.G., et al. Juvenile ossifying fibroma of the maxilla. Egypt J Ear Nose Throat Allied Sci (2016), http://dx.doi.org/10.1016/j.ejenta.2016.09.002

9. Yang HY, Zhen LW, Luo J, Yin WH, Yang HJ, Zwahlen RA. Psammomatoid juvenile cemento-ossifying fibroma of the maxilla. *J Craniofac Surg*. 2009;20:1190–1192.
10. Rosa M, Manas M, Rey MJ, Gaston F. Aggressive psammomatoid ossifying fibroma. *Ann Otol Rhinol Laryngol*. 2002;111:466–468.
11. Bendet E, Bakon M, Talmi YP, Tadmor R, Kronenberg J. Juvenile cemento-ossifying fibroma of the maxilla. *Ann Otol Rhinol Laryngol*. 1997;106:75–78.
12. Wakefield MJ, Ross AH, Damato EM, Salvi SM, Baker GRC. Review of lateral orbital wall ossifying fibroma. *Orbit*. 2010;29(6):317–320.
13. Trijolet JP, Parmentier J, Sury F, Goga D, Mejean N, Laure B. Fibrome cémento-ossifiant de la mandibule. *Annales françaises d'oto-rhino-laryngologie et de pathologie cervico-faciale*. 2011;128:34–37.
14. Singh Bist S, Varshney S, Bhagat S, Mishra S, Aggarwal V. juvenile aggressive cement-ossifying fibroma of the maxilla. *Otorhinolaryngol Clin Int J*. 2012;3:156–159.
15. Thankappan S, Nair S, Thomas V. Psammomatoid and trabecular variants of juvenile ossifying fibroma-two case reports. *Indian J Radiol Imaging*. 2009;19:116–119.
16. Banu K, Palikat SM. Juvenile trabecular ossifying fibroma of the mandible. *J Maxillofac Oral Surg*. 2010;9(1):87–90.

Please cite this article in press as: Ollfa B.G., et al. Juvenile ossifying fibroma of the maxilla. Egypt J Ear Nose Throat Allied Sci (2016), http://dx.doi.org/10.1016/j.ejenta.2016.09.002